Applicant: Tupler et al. Attorney's Docket No.: 07917-180001 / UMMC 03-18

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## Amendments to the Claims:

This listing of claims replaces all prior versions and listings of claims in the application.

## **Listing of Claims**:

- 1. (Currently Amended) A method of identifying a candidate therapeutic compound for treating [[FSHD]] <u>facioscapulohumeral muscular dystrophy (FSHD)</u>, the method comprising,
  - (a) providing a D4Z4 binding element;
  - (b) contacting the D4Z4 binding element with a test compound; [[and]]
  - (c) determining whether the test compound interacts with the D4Z4 binding element[[,]] and
- (d) selecting the test compound as a candidate therapeutic compound for treating FSHD if the test compound interacts with the D4Z4 binding element.

  wherein an interaction between the D4Z4 binding element and the test compound indicates that the test compound is a candidate therapeutic compound.
- 2. (Original) The method of claim 1, wherein the D4Z4 binding element is in a cell that expresses a 4q35 gene.
- 3. (Currently Amended) The method of claim 2, further comprising the step of determining the level of expression of a 4q35 gene in the presence of the test compound as compared to a reference representing a level of expression in the absence of the test compound, wherein a decrease in expression of the 4q35 gene indicates that the test compound interacts with the D4Z4 binding element and is a candidate therapeutic compound for treating FSHD.
- 4. (**Original**) The method of claim 3, wherein the 4q35 gene is FSHD region gene 1 (*FRG1*), FSHD region gene 2 (*FRG2*), or adenine nucleotide translocator-1 gene (*ANT1*).

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5. (Currently amended) The method of claim [[1]] 2, wherein the cell is a muscle cell.

- 6. (Original) The method of claim 5, wherein the cell is from a subject that has FSHD.
- 7. (Original) The method of claim 1, wherein the interaction is the binding of the test compound to the D4Z4 binding element.

## 8. - 9. (Cancelled)

- 10. (Currently Amended) A method of identifying a candidate compound for treating FSHD, the The method of claim 1, further comprising,
  - (a) providing a D4Z4 binding element (DBE) and a D4Z4 recognition complex (DRC) under conditions such that the DBE and the DRC can interact bind to each other;
  - (b) contacting the D4Z4 binding element and DRC or a DRC component with a test compound identified by the method of claim 1; and
  - (c) determining whether the test compound affects the interaction between binding of the D4Z4 binding element and to the DRC or DRC component,

wherein an increase in the interaction binding between the D4Z4 binding element and the DRC or DRC component in the presence of the test compound indicates that the test compound is a candidate compound.

11. (Currently amended) The method of claim [[9]] 10, wherein the DRC component is [[YY1]] YinYang 1 (YY1), HMGB2 High Mobility Group Box 2 (HMGB2), or nucleolin.

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12. - 20. (Cancelled)

21. (New) The method of claim 1, wherein the D4Z4 binding element is in a reporter construct comprising a promoter and a reporter gene.

- 22. (New) The method of claim 21, wherein the reporter construct is in a cell, and the method comprises detecting a level of expression of the reporter construct in the presence of the test compound as compared to a reference representing a level of expression in the absence of the test compound, wherein a decrease in expression of the reporter construct indicates that the test compound interacts with the D4Z4 binding element and is a candidate therapeutic compound for treating FSHD.
- 23. (New) The method of claim 21, wherein the reporter construct comprises one, two or six minimal D4Z4 binding elements.